Multiple dural-based hemangiopericytomas

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Here we report the case of a 57-year-old man who underwent resection of a dural-based hemangiopericytoma (HPC) in the left frontoparietal region. The patient was treated with radiation therapy and remained symptom-free for 10 years. At 67 years of age, he presented with a mass in the left frontal region near the same area as the first tumor, in addition to a separate smaller mass in the right middle cranial fossa. Resection of the larger left frontal mass revealed an HPC. Follow-up imaging 9 months later showed a significant increase in size of the right middle cranial fossa mass. This third mass was resected, and histological examination also demonstrated an HPC.

Hemangiopericytoma (HPC) was first described in 1942 by Stout and Murray as a highly vascularized tumor arising from the pericytes of Zimmerman, the modified smooth muscle cells that surround capillaries and post-capillary venules (1). In 1954, Begg and Garret first reported a case of an HPC occurring in the nervous system (2). These are rare neoplasms representing 2.5% of all meningeal tumors and 0.4% of all intracranial tumors (3). The 2007 World Health Organization (WHO) criteria developed a two-tiered system for grading intracranial HPCs, including WHO grade II and grade III (anaplastic) (Table 1) (4). In the largest published review of meningeal HPC, including follow-up data on 277 patients, local recurrence was seen in 57% of patients and systemic metastases was seen in 27% (5). Isolated intracranial metastasis remains an unusual event. The overall literature on intracranial HPC remains limited, especially concerning long-term follow-up of patients (6, 7).

CASE REPORT

A 57-year-old man without any significant family history or past medical history underwent resection of an extra-axial mass in the left frontoparietal region. The pathologic diagnosis of this first tumor was HPC, WHO grade III. The patient was treated with radiation therapy and remained symptom-free for approximately 10 years.

When the patient was 67 years of age, radiologic imaging showed an enhancing partially cystic extra-axial mass in the left frontal region (Figures 1a and 1b). Another smaller mass was also visualized in the right middle cranial fossa (Figure 1c). He

Table 1. 2007 World Health Organization two-tiered criteria for intracranial hemangiopericytoma

Grade	Features
II	Monomorphous cellular spindle-cell tumor with round-to-oval nuclei and intratumoral staghorn vessels
III (anaplastic)	Necrosis and/or Mitotic activity (<5 per 10 high-power fields) <i>Plus at least two of the following:</i> —Hemorrhage —Moderate to high cell density —Moderate to marked nuclear atypia

underwent resection of the larger left frontal mass, and histological examination revealed an HPC, WHO grade III. Follow-up imaging 9 months later revealed a significant increase in size of the right middle cranial fossa mass (Figure 1d), which did not appear to have obvious connection to the previous two masses. The decision was made to proceed with resection of this third mass due to its rapid growth rate. Histological examination again revealed an HPC, WHO grade III. Subsequent imaging ruled out systemic metastases. At 1 year postresection, the patient does not have any additional symptoms and continues to be an active rancher.

The original left frontoparietal mass (the first of the three tumors) was highly cellular with a very rich vascular network (Figure 2a). The vascular channels displayed a staghorn-like appearance. Multifocal necrosis and hemorrhage were identified. The mitotic activity was up to 5 mitotic figures per 10 high-power fields. Immunohistochemical studies showed prominent

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Figure 1. Ten years after the first mass was resected, the patient presented with worsening neurological symptoms. **(a)** An axial contrast-enhanced T1-weighted image showed a left frontal, extra-axial, avidly enhancing, partially cystic tumor resulting in significant mass effect on the left frontal lobe and lateral ventricles. **(b)** An axial T2-weighted image demonstrated the extent of vasogenic edema in the frontal lobes and genu of the corpus callosum. The dark signal in the cystic portion of the mass posteriorly represents dependently layering hemorrhage. **(c)** In addition, axial contrast-enhanced T1-weighted imaging showed a right middle cranial fossa, extra-axial, avidly enhancing mass distant from the left frontal mass. **(d)** An axial contrast-enhanced T1-weighted image acquired 9 months later showed the right middle cranial fossa mass to have increased in size.

CD34 reactivity and negative epithelial membrane antigen. The morphology and immunostaining patterns supported a diagnosis of an HPC, WHO grade III.

The patient's mass in the left frontal region, which was removed 10 years later, showed histopathology characteristics similar to those of the first tumor (*Figure 2b*). Reticulin staining highlighted the rich microvascular network (*Figure 2c*). As with the original tumor, CD34 reactivity was diffusely positive and epithelial membrane antigen was negative. The morphology and

staining patterns were again diagnostic of an HPC, WHO grade III.

The right middle fossa mass was resected 9 months after the second left frontal mass and was microscopically similar to the previous tumors (*Figures 2d and 2e*). Mitotic activity was focally brisk with up to 5 mitotic figures per 10 high-power fields. There was multifocal coagulative necrosis (*Figure 2f*). Immunohistochemistry showed tumor cells to be diffusely and strongly immunoreactive for CD34 (*Figure 2g*) and CD99 (*Figure 2h*), whereas keratin, epithelial membrane antigen, and smooth muscle actin were negative. These findings were again diagnostic of an HPC, WHO grade III.

DISCUSSION

The multifocal presentation of intracranial HPCs can have three explanations: 1) recurrence of a previously resected HPC; 2) metastasis of an HPC to other intracranial, dural-based locations; or 3) development of independent HPCs at different sites. In the present case, it is most likely that the second tumor was a recurrence of the primary tumor resected 10 years earlier since the second HPC arose at a site very close to the first HPC (left frontoparietal region). The third dural-based HPC, however, developed in an entirely separate area of the brain (in the right middle fossa), relatively distant from the site of the first two HPCs. There is no specific vascular connection favoring this particular spread; however, there is a possible, although distant, cerebrospinal fluid connection. It is less likely that the third HPC developed independently of the first two HPCs. In most cases of intracranial HPC with reported recurrence

and/or metastases, recurrence at the primary tumor site is usually followed by systemic rather than intracranial metastases.

The interesting feature of this particular case is the lack of systemic metastases in the setting of an unusual intracranial metastasis from one dural-based location to another without an obvious connection. Since recurrence with a greater degree of malignancy can develop following an extended disease-free interval, this case highlights the importance of careful long-term follow-up for patients with HPC.

Figure 2. (a) Hematoxylin and eosin (H&E)-stained section of the original tumor showing a rich vascular network with a staghorn-like appearance. (b) H&E-stained section of the second tumor removed 10 years later showing patternless sheets of plump cells with indistinct cell borders and abundant vasculature with a staghorn-like appearance. (c) Reticulin stain of the second tumor, highlighting the rich microvascular network and pericellular reticulin fibers. (d–f) H&E-stained section of the third tumor 9 months later, showing a highly cellular tumor composed of sheets of cells in a highly vascular background with (d, e) a vaguely staghorn-like configuration and (f) multifocal coagulative necrosis. Immunohistochemistry showed tumor cells to be diffusely and strongly immunoreactive for (g) CD34 and (h) CD99.

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